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PROGRESSIVE MUSCULAR ATROPHY; A STUDY OF THE
CAUSES AND CLASSIFICATIONS, WITH THE
REPORT OF AN AUTOPSY.*

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OF NEW YORK.

The present paper is a clinical study of the records of 72 cases of progressive muscular atrophy, of bulbar or spinal origin. It includes none of the muscular dystrophies, but only those forms of muscular atrophy which are progressive in character, and are degenerative and central in origin, viz., progressive ophthalmoplegia, bulbar paralysis, amyotrophic lateral sclerosis, and the various types of spinal progressive atrophy whether beginning in the arms, legs, shoulders, or hip girdle. I have added the record of one case of progressive bulbo-spinal palsy and tabes, with autopsy.

There are three points which I have studied especially, in connection with these histories, and which I wish to present for discussion:

1st. The clinical classification of these cases.

2nd. The etiology and, especially, etiological relation of syphilis to this disease.

3rd. The question of the proper status of so-called "chronic anterior poliomyelitis," and its relation to the atrophies.

CLASSIFICATION.—As regards the classification, the status of *amyotrophic lateral sclerosis* has, perhaps, caused the most discussion, in the past, amongst neurologists. In my list I have histories of only three cases of progressive muscular atrophy, be-

* Read at the meeting of the American Neurological Association, June 2, 1905.

ginning with symptoms of spasticity, and accompanied by this condition very dominantly, throughout its course, with all the objective evidences of sclerosis of the lateral columns. These cases, while undoubtedly closely related, and pathologically identical with other types, have such distinct clinical features, that the use of a special term to designate them, seems to be sufficiently justifiable. If, however, I add to this list of three, all the cases of progressive muscular atrophy which were later associated with *some signs* of spasticity, and *some evidence* of the lateral cord involvement, the number would be 14. Of these 14, four began with symptoms of the ordinary type of progressive bulbar paralysis, and only after some months developed spastic symptoms. Two others began in the legs and ascended to the arms, shoulders and bulb, developing late spastic symptoms, while five more began in the hands and arms, simulating at first, perfectly, the ordinary type of the Duchenne-Aran atrophy, but later appearing like cases of amyotrophic lateral sclerosis.

Thus, cases beginning in the same way progress, some with and some without developing spasticity. It seems, therefore, a rather unnecessary confusion of symptomatology to transfer the diagnosis of progressive muscular atrophy, of the Duchenne-Aran, or other type, to that of amyotrophic lateral sclerosis, as soon as a little evidence of spasticity begins to show itself, especially as there was nothing peculiar either in age, course or duration. In one patient, for example, the atrophy began in the hands, and ascended to the shoulders, when there was some evidence of lateral cord involvement. The disease was then arrested, the spastic symptoms disappeared, and the patient now presents a type of Duchenne-Aran atrophy, without any spastic symptoms. I should therefore, I repeat, limit the use of the term, "amyotrophic lateral sclerosis," to those cases which show only from the beginning and dominantly, the spastic and contracturing types of progressive muscular atrophy.

Progressive Bulbar Palsy and Progressive Ophthalmoplegia.—

There is a very close similarity in the etiology, course and termination of those cases which begin in the arms and then ascend to the medulla, and those cases which begin in the medulla or ocular nuclei and then descend and involve the hand, arm and legs. I have found that all my cases of progressive bulbar paralysis and

ophthalmoplegia, if they lived long enough, showed, eventually, signs of a spinal muscular atrophy also.

Thus I have the histories of two cases of progressive ophthalmoplegia; the disease showed a descending course, involving the mid-brain nuclei, then the medulla, and then the spinal centres, until the whole of the oculo-motor, bulbar and spinal motor neurones were involved. One-half the cases of progressive bulbar palsy, starting in a most typical manner, with dysphagia, dysarthria, developed eventually, or almost simultaneously, symptoms of involvement of the arms and legs, and sometimes, associated spastic symptoms. I have now a patient who has in the last six months slowly developed a glosso-laryngeal atrophic palsy and left-hand atrophy without spasticity. I see no reason, therefore, for not including under one name, all forms of progressive muscular atrophy, and separating them, if we must, for purposes of convenience, into those types which begin in the eyes, the bulb, the cervical or the lumbo-sacral cord. According to this method, I find amongst 72 cases, three of amyotrophic lateral sclerosis, there being over 55 of the other form. Of this 55, eleven began in the mid-brain or bulb and descended, while 33 began in the arms, and 4 in the legs and ascended.

It will be seen that the cervico-bulbar (or Duchenne-Aran) type of progressive muscular atrophy, far exceeds in frequency, any other form, and a certain amount of spasticity develops in these, in about 25 per cent. of the cases.¹

¹ There is a curious similarity between the methods of onset in the whole group of cases, including bulbar paralysis and progressive muscular atrophy, on the one hand, and in those of amyotrophic lateral sclerosis alone, on the other. Thus, in a list of 81 cases of the latter disease, collected by Collins, the upper extremity was affected in 39 cases, or about 50 per cent. the lower extremity in 14 cases, or about 12 per cent.; hemiplegic and all four extremities in 11 cases or about 8 per cent., and in the bulb in 21 cases, or about 25 per cent.

In 40 cases of my own, the disease began in the upper extremities, in 17 cases, the lower extremities, in 7 cases, and in the four extremities and hemiplegically, in 13 cases, and in the bulb, in 13 cases.

DANA.		COLLINS.	
ALL TYPES.		AMYOTROPHIC.	
Bulbar and eye	13	Bulbar	21
Cervico-spinal	17	Cervico-spinal	39
Lumbo-spinal	7	Lumbo-spinal	11
Quadruplegic and hemiplegic	13	Quadruplegic and hemiplegic ..	11
	40		82

The attempts to group the cases as those of amyotrophic lateral

So far, therefore, I make out essentially only one kind of progressive muscular atrophy,—and this includes amyotrophic lateral sclerosis, ophthalmo, bulbar and spinal progressive palsies.

Progressive Occupation Atrophies.—There is, however, a group of cases, which seem to me to run a somewhat special course. They are those which begin in the shoulder girdle, and then pass to the hip girdle, or *vice-versa*. Then they may slowly extend to the trunk and extremities. They occur in rather young people, and in almost all cases can be attributed to occupation, combined, perhaps, with some excesses that otherwise deteriorate the constitution. They run a much longer course than the other forms of atrophy, and there occur periods of arrest. They show very marked fibrillary twitchings, and the usual absence of degenerative reactions, until late in the disease. They are not associated with any lipomatosis, nor any family history of atrophy, nor history of syphilis, nor are there in connection with their atrophies, any of those peculiar vascular and trophic disturbances, which are usually associated with the progressive muscular dystrophies. They give no evidence of sensory trouble or those degenerative reactions which occur in progressive atrophies of peripheral, nervous origin.

I have no sections which will prove that they were not dystrophies, however, and in their slow course and relatively benign progress, they bear some relation to them. They may be, perhaps, simply classed as “progressive occupation atrophies” that have become extensively spread, and owing to a weakened or defective constitution, they acquire a slowly progressive character. Their clinical recognition and differentiation is justified on the ground of their better prognosis. One of these cases which occurred in the person of a professional contortionist, will illustrate the character of the trouble better than any general description.

sclerosis on the one hand and as cases of progressive spinal muscular atrophy on the other was found impossible, for the cases with spastic symptoms begin and run their course in very different ways.

Thus, of 15 cases beginning with bulbar or eye and bulbar symptoms, only 2 developed later typical spastic symptoms like those of amyotrophic lateral sclerosis.

Of 23 cases beginning in the arms like a Duchenne-Aran type, six developed spastic symptoms later.

Of 12 cases beginning in the shoulders or thighs and shoulders, 4 developed spastic symptoms.

Of 12 beginning in the legs, or hemiplegically, 2 developed spastic symptoms.

PROGRESSIVE OCCUPATION MUSCULAR ATROPHY.

James De L., aged 27, white, Irish, admitted to Bellevue Hospital, March 22, '97.

Family History: Mother died of consumption—otherwise negative; has four brothers and three sisters, all well.

Previous History: He cannot recall being sick at any time previous to the present trouble. His father was a horse-trainer in a circus, and he was brought up to be a contortionist as far back as he can remember. He had a brother who was a contortionist, and was perfectly well up to the latest information. He had no syphilis, no gonorrhea previous to the present disease; but he did drink from one to two pints of whiskey daily for a period of about five years.

Present Illness: In Nov., 1886, while performing, he fell from a trapeze and injured his back. This was followed by great weakness and pain in the back. There were no spinal pressure symptoms, and he otherwise felt all right. He went to St. George's Hospital, London, and remained in bed ten weeks. Four weeks after his entrance a plaster of Paris jacket was applied and extension to the limbs was given as they "jerked up" a good deal. The jacket he wore four months in the country where he went to grow stronger. The jacket was then removed and he returned to his profession, feeling as strong as ever. He remained perfectly well and performed his duties as usual for four months. One night as he was going to perform he noticed his former pain in the back and found he could not do his work. This was nine years ago at the age of 18. He has not been able to do anything since. The history since that night is as follows: Pain in back continued for five or six years; it was worse on exercising, but was scarcely felt on resting. Both the shoulders began to atrophy in four months, followed soon by the arms and forearms. These atrophied to their present condition, in about 12 months, then stopped. Following this he had three years of respite, then he noticed his waist growing thin; this was soon followed by thinness of calves. He was able to walk, however, up to 12 months ago; since then he has been in a wheel-chair. His general condition during this time has been good, except for periods of severe headaches, lasting three days sometimes and associated with blurred vision, swelling of eyelids and nausea and vomiting. He had no other sensory symptoms. He came to America two years ago, and three months after came to Bellevue, where he remained one and a quarter years.

Physical Examination.—Patient is poorly nourished. Tongue coated slightly. Subcutaneous fat seems present in good quantity, skin soft and moist. Arms, thighs and waist markedly wasted. Lungs negative. Heart action good. Loud systolic murmur heard over base and transmitted upward. Liver and spleen negative.



Fig. 1.

Nervous System.—Mental and cerebral symptoms absent.

Sensory—touch, temperature, space, pain, location, position, form, normal. No anesthesia, hyperesthesia or paresthesia.

Motor.—Muscles of forearms, calves, buttocks, trunk, face and neck firm, but small and atrophied, though he has some voluntary motion in all except the *erector spinæ*. There are fibrillary twitchings in the muscles of thigh and calf. The rhomboidei are markedly atrophied, giving winged appearance to scapulæ. Pectorals are much atrophied, so that ribs in front are very prominent. There is a marked anterior-posterior curvature of spine, convexity forward, in the lumbar and lower dorsal region. Respiration is normal, and he does not use accessory muscles. Sphincters normal. Face and masticatory muscles normal. Muscles of hand not atrophied. Has some talipes equino-varus. Although patient cannot stand, he still manages to dress, etc., without any assistance, making one hand help the other, etc.

Electrical Examination.—Forearms, lessened galvanic reaction. Arms, thighs, calves, rhomboids show degeneration reaction. Face and hand muscles normal.

Comparative Measures.

Left arm, 2 in. above elbow	Forearm, 2 in. below elbow.
Right 5¼ in. circum.	7½ in.
Left Thigh, 4 in. above knee.	Calf, 4 in. below knee
Right 9 in.	10 in.
Right 5½ in. circum.	10½ in.

This shows the arms and thighs to be smaller by ½ in. than the forearms or legs. It is a so-to-speak "root-joint" atrophy.

Vaso-Motor.—Lower part of legs cold and bluish; no trophic disturbance except in muscles.

The disease slowly progressed, and the patient finally died of an intercurrent disease without involvement of medulla. Total duration of disease about eleven years.

Chronic Anterior Poliomyelitis.—There is another type of progressive muscular atrophy which deserves, I think, more attention from American neurologists than it has received, viz., that, characterized by what we might suppose to be a chronic anterior poliomyelitis.

There seems to be a great difference of opinion amongst neurological writers, as to the actual existence of any such disease as this. I have, myself, seen and reported one case in which the lesion was confirmed by autopsy (loc. cit.), but it was a subacute rather than chronic disorder. Starr, in his recent work, puts progressive muscular atrophy under the head of chronic anterior poliomyelitis, while I have simply classed under this name, the

chronic cases of acute anterior poliomyelitis, not thinking one justified in recognizing as a special disease the subacute and chronic poliomyelitis of Duchenne.

The literature of the subject, as given in the "Handbuch der pathologischen Anatomie des Nervensystem," is limited to 11 references. To this may be added a case of my own, reported with Dr. Biggs (JOURNAL OF NERVOUS AND MENTAL DISEASE, June, 1887), male, 53, gradually atrophic paralysis, of six months' duration, ending in death. Central myelitis with cavity. Also one reported by J. B. Charcot "L'atrophie musculaire progr.," 1895.² p. 115. Nonne, in 1891, analyzed previous cases, 5 in all, and made out of these five, three types:

1. Circumscribed, reaching a certain stage and remaining stationary (Eisenlohr's).⁴
2. With rapid atrophy and paralysis, with tendency to regression and cure (Landouzy-Dejerine).³
3. A chronic, steadily progressing, atrophic paralysis, with or without involvement of peripheral nerves (Dreschfield, Oppenheim, Nonne).⁵

In Nonne's case and Oppenheim's there was a progressive degeneration of the anterior corneal cells and a slight involvement of the lateral white columns, but *no inflammatory reaction*.

In none of these cases, except perhaps Eisenlohr's and my own, was there any inflammation, and the condition was distinguished from a progressive atrophy by its more rapid course (about two years), and the fact that atrophy preceded the paralysis.

The situation, as regards chronic and subacute anterior poliomyelitis, then is about the following:

1. Some cases reported as such, in the past, were cases of multiple neuritis.
2. Other cases were those of a subacute, atrophic paralysis, running a course of about two years or less, and due to a rather rapid cellular degeneration, not to inflammation. In other words,

²In Charcot's case there was no history of syphilis, but the autopsy showed marked vascular lesions. He states that a sclerosis of the anterior fundamental column is characteristic of this disease.

³Landouzy-Dejerine, Rev. de Méd., 1882, No. 2, 12.

⁴Eisenlohr, Neurolog. Centralbl., 1882, No. 18.

⁵Oppenheim, Archiv. f. Psych. xix, Hft. 2, 1888.

⁵Dreschfield, Brain, 1885, July.

⁵Nonne, Deut. Zeit. für Nervenheilkunde, Vol. 1, p. 138, 1891.

they were really cases of progressive atrophies, so that this rapid course is not a diagnostic criterion.

3. There are, however, some clinical cases (none as yet with autopsy so far as I know), in which paralysis and atrophy affect certain cell groups, in a relatively active fashion, i.e., in a few weeks. The disease then stops, and later another group of nerve cells is attacked in the same way, and later a third or fourth. We have in these a clinical picture, which corresponds to a subacute poliomyelitis. They are probably cases of *cellular degeneration from vascular disease (syphilitic?)*.

There is then, I conclude, not yet established on a sure pathological basis, such a disease as a true chronic or subacute inflammation of the anterior horn-cells of the spinal cord. The cases so reported, are instances rather of subacute degenerative processes, secondary sometimes to vascular disease, and are probably related to the progressive, spinal atrophies. Degenerative change may occur rapidly and destroy nerve cells in a few months as well as in five or six years, and this fact explains the reports of clinical cases, diagnosticated as "chronic poliomyelitis anterior."

These *subacute progressive atrophies*, however, should be recognized clinically, and it is probable that the process is due to vascular disease primarily, and is of specific origin.

I have two cases illustrating this kind of "*subacute, remittent spinal atrophy*." In one case the patient had a gradual development, in two or three weeks, of a paralysis of one leg, associated with some irritative symptoms, leaving the leg weak and atrophied. After some months or a year, the arm on the same side became affected, then the other arm, and then the other leg, finally leaving him in a condition of chronic atrophic paralysis. In the other case the patient became affected with a subacute motor paralysis of the shoulders, developed in the course of two or three months; this became arrested, and then after a year, another series of segments of the spinal cord, higher in the neck, became affected in the same way, leaving the patient in a few weeks with a further development of his atrophic paralysis. It was, like the other, a remittent, progressive atrophic paralysis.

One of these cases was presented to the New York Neuro-

logical Society, by Dr. Hunt, as representing a form of subacute poliomyelitis anterior.

These patients showed fibrillary twitchings, they developed spastic symptoms, and they have all the general symptoms of ordinary progressive muscular atrophy, but the paralysis was, at least, a little in advance of the muscular atrophy, and there were evidences of the involvement of other parts of the spinal cord than the anterior horns,—such as the lateral fundamental columns and the pyramidal tracts.

In both my two cases of this remittent disorder, there was a very unmistakable history of syphilitic infection. I think it likely that some of the other cases which have come under my observation, if it had been possible to watch them closely for a long time, would have shown a progress somewhat similar to this. So that, it seems to me, apart from the true progressive spinal atrophies, there may be yet developed a type of *recurrent subacute poliomyelacea*, which should be separated from the ordinary groups, and it will further be found that *syphilis* is perhaps the infection which is the cause of this form of atrophic paralysis.

I append summaries of the histories of these cases:

Subacute Progressive Atrophy, coming on in successive attacks.

Case I. H. M., male, age 40, English, boiler-maker, lues in 1889, with secondaries, and but little treatment. No alcoholism or lead. In January, 1901, weakness, stiffness and atrophy in left upper arm and shoulder. In August, 1902, same process in the right arm and shoulder. Admitted to the hospital at that time, with atrophic paralysis of both shoulders and upper arms, the parts involved being the biceps, brachiales antices, deltoid, spinati, supinators and pectorals,—the left side worse than the right; exaggerated knee-jerks, ankle-clonus and Babinski. Fibrillary tremors. Patient's condition remained stationary for about a year. In February, 1904, the same process began to involve the extensors of the neck, so that the chin fell on the chest and could not be raised (Fig. 2). At the present time he has atrophic paralysis of upper arms, shoulders, and posterior neck muscles, with slight exaggeration of reflexes; no ankle-clonus. Pupils are of the Argyll-Robertson type. He has had no anesthesia or pain, but has had slight feelings of numbness in shoulders and arms. Patient's condition has since second attack remained nearly stationary for about a year.

Case II. P. D., male, 39, single, waiter, Irish, alcoholic excesses; lues in 1892 and secondaries. In 1895 began to lose power in right leg with atrophy. In 1896 same process in right arm and shoulder. After an interval of several months, the left arm and

shoulder, and after another interval of several months, the left leg and then trunk muscles became involved. Finally, in 1899, four years after the attack began, the medulla became affected. The disease began each time with a sense of weakness, some fibrillary twitchings, and crampy, painful sensations. Paralysis and atrophy followed. The process recurred at intervals, until in two years all

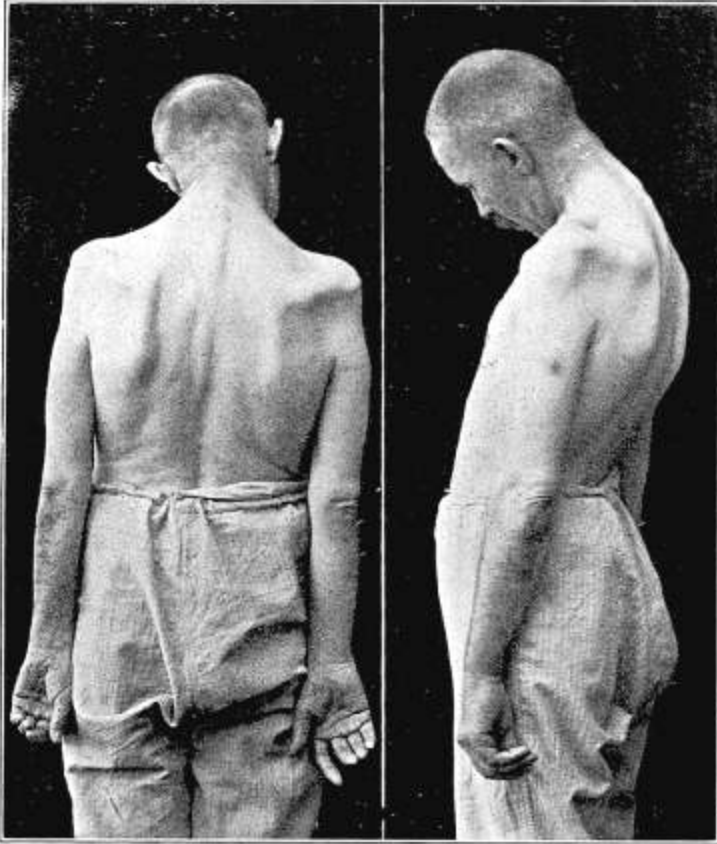


Fig. 2. Case of Subacute Remittent Progressive Muscular Atrophy. Patient is unable to raise the head.

four extremities were involved, and in four years the trunk muscles and bulb.

We have then three kinds of progressive muscular atrophy of central origin:

1. The Bulbo-spinal type including ophthalmoplegia, bulbar

palsy, cervico-spinal (Duchenne-Aran) atrophy and the various leg types.

2. Progressive occupation atrophy, slower in course with a tendency to affect the shoulders, trunk and hip-girdle more than the legs and forearms.

3. Subacute anterior poliomyelacea coming on usually in remittent attacks and probably of specific origin.

ETIOLOGY.

(1). *Syphilis and Progressive Muscular Atrophy*.—With regard to the relation of syphilis to progressive muscular atrophy, the facts that I have to furnish can soon be given, and I believe that they are, at least, more than suggestive. I have seen so many cases of progressive muscular atrophy that were so distinctly parasyphilitic, that it seemed to me we might infer that nearly all the atrophies were due to this process. Amongst my 72 cases, 19 gave a history of syphilis, which would give a percentage of about 25 per cent. Leaving out the shoulder-hip type, which seemed to belong to a different category of disease, the presence of syphilis as the cause of progressive muscular atrophy of all forms, would be greater and probably if the antecedent history were more definite in all cases, the percentage would receive another increase. It will be remembered that the investigation into the syphilitic origin of tabes, began with rather modest statistics in its support. In none of my three very distinctive cases of amyotrophic lateral sclerosis, was there a history of syphilis. On the other hand, in 33 cases, beginning in the arm and hand, and of the ordinary Duchenne-Aran type, there were 9 cases. Among the 14 cases, associated with spastic symptoms, there were 3 cases of syphilis, and in the bulbar or bulbo-cervical type, of which there were 11, there were 3 cases, while in 5 of the hemiplegic type, there were two cases.

In all types, therefore, whether beginning in arms, legs or medulla, the percentage of syphilis seems to range about the same, which would be another confirmation of their pathological unity. Of the parasyphilitic cases, three were associated with the early symptoms of tabes, and one with symptoms of paresis. In 2 cases, persons who had had an old infection of syphilis, had developed atrophies in the arm and hand, of a very slowly progressive type. The late and degenerative influence of the syphil-

itic poison, may therefore show itself primarily in the form of a progressive atrophy of a perfectly typical kind, or it may show itself in the form of a locomotor ataxia, with a mild grade of progressive muscular atrophy, slowly progressing, and imposed upon the tabes.*

Since writing up my statistics, I have read the paper of Lari (*Arch. de Neurologie*, Vol. XVI, 303) who reports 30 cases of amyotrophic lateral sclerosis with syphilitic history and cites the case of a physician who suffered from amyotrophic lateral sclerosis, and who was cured by antisyphilitic treatment. It may be that the same vigorous early treatment of progressive muscular atrophy on anti-syphilitic lines, will help to arrest some cases of this dread disease.

I append here a case of this kind:

Case of Tabes and Bulbar Spinal Palsy.

L. B., age 60. French, teacher. Admitted March 30, 1903. Mother died of acute phthisis, otherwise comes of long-lived, healthy stock. Moderate use of wine, beer, cognac and absinthe; four to six cigars daily. 40 years ago epididymitis-gonorrhea. One year later (39 years ago) had a sore on penis—cauterized; no secondaries, no constitutional treatment. 25 years ago acute articular rheumatism. 22 years ago jaundice, probably catarrhal. Has been an uncommonly healthy and active man, and a great talker.

Present Illness.—Six months ago his customary walks caused him fatigue and pain in back; this was quite severe, but was relieved at once on sitting down. Exertion also produced dyspnea. At the same time, difficulty in enunciating distinctly, developed especially in pronouncing *l, r, n*; a little later there was trouble in masticating and swallowing large pieces of solid food, and his throat felt narrower than usual. He soon noticed weakness in the upper extremities—the small muscles of the hands being first

* Among 19 cases in which there was a history of syphilis the patients were all males, and ranged in age between 33 and 60, most being about 40. There were in some a contributing history of alcohol, tobacco, overwork, or trauma. In 4 the disease was associated with tabes, and of those 4, one began as a bulbar palsy, the trouble extending through to the spinal cord. In the other three the disease began in the arms like a Duchenne-Aran type; in two of these the process was arrested, the luetic infection being a very old one.

In 8 the disease became associated with spasticity and exaggerated reflexes and clonus. In one it began in the shoulder girdle, in one in the legs, in the other 5 it began in the hands and forearms, and in all but 1, eventually involved the medulla.

In 2 cases the disease came on in the leg, arm and shoulders of one side and in 1 case remained hemiplegic in type.

In one case a patient after an infection had tabes for 20 years, then developed bulbar palsy and died.

affected, so that delicate movements, as shaving and writing, became difficult. The extensors of the forearm and hips also became weak and atrophic. He had difficulty in walking. Vertigo occurred on rising suddenly. On admission to the hospital he complained of pain in back, dyspnea was so great on exertion that he could hardly walk a block. Dysarthria was marked, and he had to use semi-solid or liquid nourishment. Smell, taste and vision normal; *g, l, r, x*, difficult to enunciate. Motor weakness—atrophy of shoulder girdle—fibrillary tremor present. Legs showed no marked atrophy. There was diminution of pain and touch sense in the fingers. The wrist and triceps jerks were absent; the knee jerks and ankle jerks were very feeble.

No Babinski. No Romberg. Sphincters normal. Argyll-Robertson pupil present. He suffered from characteristic darting pains in legs. Pulse temp. and resp. showed nothing of note.

Later notes, by Dr. J. R. Hunt.—Patient is a tall, round-shouldered and slightly-stooped, but healthy-looking old man, with white hair and ruddy complexion. He walks with marked eversion of feet (due to old Potts' fracture of right leg.) Speech is thick and articulation indistinct, with marked laryngeal hoarseness. For five years hardness of hearing (slight) on both sides, but this has not increased noticeably during present illness. He has at times a distinct girdle pain. Pulse 75, regular, good quality, no tension, no thickening. Heart, no murmurs, no accent. Lungs moderate, general bronchitis.

Cranial Nerves.—3, 4, 5, 6, and 7 nerves are normal. The orbicularis oris is defective, noticed in forming labials; cannot whistle as formerly or blow out a candle.

On innervation of the palate a weakness is quite apparent. It is not lifted in normal fashion—the movement is better on the left side. Uvula is practically normal. On protruding tongue, a marked atrophy is apparent, the surface is wrinkled and corrugated, and the seat of constant fibrillary twitchings. The larynx is lifted readily during the act of deglutition. No respiratory and no cardiac symptoms. There is atrophy and weakness of the muscles of the shoulder girdle—is unable to lift arms high above the head. The muscles of the upper arm and forearm are fairly strong. The hands are weak and the thenar eminences appear slightly atrophied. Power of legs about normal—no atrophies. The knee-jerks and Achilles jerks are lost. Triceps jerks present, but not exaggerated. The jaw-jerk is active. The right pupil is wider than the left—stiff to light and accommodation. No vesical symptoms. Very slight Romberg. Small areas of analgesia over legs.

The patient's condition at first improved. He then grew steadily worse, the bulbar symptoms predominating and he died at the end of four months, his disease lasting less than a year.

Autopsy and Histological Examination by Dr. Hunt.

Brain.—The arteries of the circle of Willis are practically

normal. The basilar shows arterio-sclerosis and the walls are thickened, but nowhere occluded. The middle cerebrals on both sides are the seat of well marked arterio-sclerotic changes (not occluded.) The roots of the hypoglossal nerves are distinctly smaller and thinner than normal (atrophic.) Otherwise, the inspection of the brain was negative. The pons and medulla were cut in series of sections, and each alternate section treated in alcohol (Nissl.) and Müller (Weigert.)

Microscopical.—Atrophy and degenerative changes are present in the 12th nerve nuclei and the motor nucleus of the glosso-

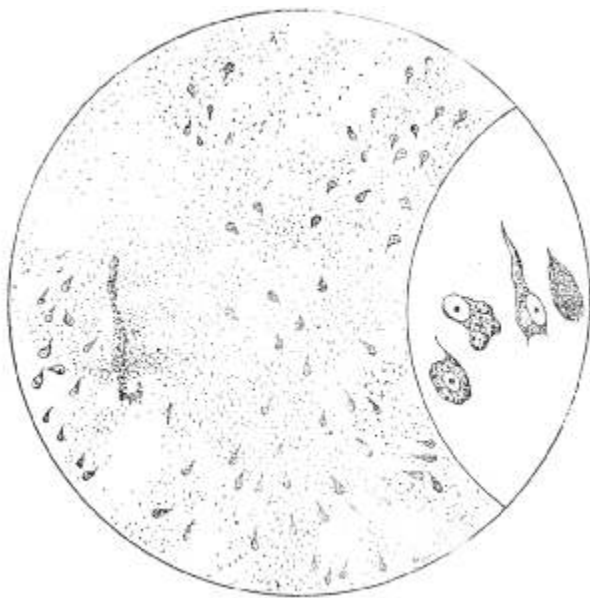


Fig. 3. Bulbar palsy and tabes, nucleus of hypoglossal. The dark cells are nearly obliterated with pigment, x2-3. To the right are four cells in different stages of degeneration, x1-6.

pharyngeal-vagus. The changes are more marked, and more striking in the 12th nerve nuclei.

Types of cells and degenerations noted were:

I. Atrophy without distinct cell destruction, the staining being intense.

II. Hyaline appearance, structureless. Rounded outline, no processes or internal structure apparent.

III. Fragments, the outline and general arrangement of which alone indicate its cellular origin.

IV. Cell rounded—no process, nucleus, protoplasm and pigment present.

V. Shrunken and vacuolated cells.

No changes noted in the other nuclei of the medulla or pons.

Weigert-Pal. The pyramidal tracts, the ascending roots of the trigeminus and the glosso-pharyngeal are normal. The root fibres of the hypoglossal nerves are atrophic, and to less extent those of the glosso-pharyngeal, which are pale and thin. The blood vessels of the medulla and pons are moderately thickened, no obliterations. The meninges moderately thickened. No foci of round-cell infiltration or other evidences of inflammatory change.

In the lower portion of the medulla as the posterior

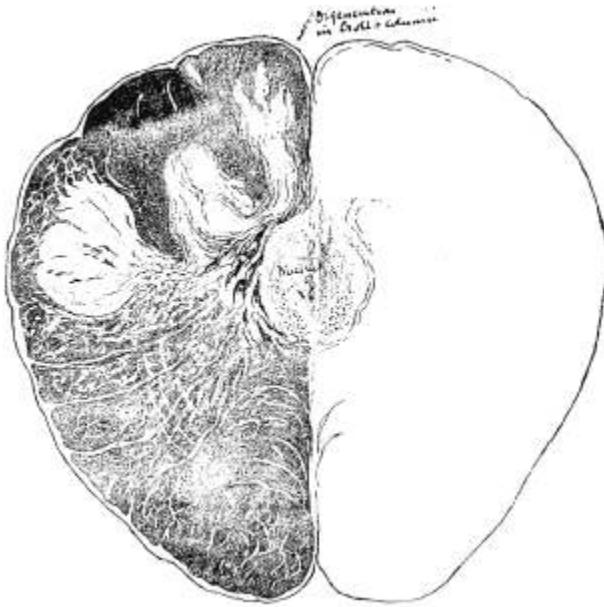


Fig. 4. Section of medulla in cases of bulbar palsy with tabes. The section is very normal except for the cell atrophy.

columns enter the nuclei of Goll and Burdach an *apparent* paling of the column of Goll is present posterior to the nucleus of Goll, by the Weigert-Pal method (Fig. 4). The neuroglia is increased in this region, some fibres lack a myelin sheath, and some are represented by empty sheaths. These changes are not present in the columns of Burdach and may represent mild tabetic changes, ascending degenerations in the columns of Goll, although the fibres of this column are noticeably small and the *glia proportionately* increased.

GENERAL ETIOLOGY.

With regard to other points in the etiology, my statistics correspond, in general, with those that have been made by others.

As regards race, it has seemed peculiar that there should be so few instances of the spinar form of atrophy in the Hebrew race, in which race there are so many cases of the dystrophies, I found no records of this malady, in the Montefiore Home, during the last fifteen years.

With regard to age, I have seen two cases beginning as early as the twelfth year, of the Duchenne-Aran variety, in the most characteristic form. In one case, which I have now watched from the time it began, at the twelfth year, to the present time, the patient now being 24, the disease was arrested, after about four years of progress, the patient being left with simply an atrophy of the left forearm, in the muscles of the ulnar distribution. My oldest patient was one of a perfectly typical arm type of atrophy, seen with Dr. J. Arthur Booth, occurring in a physician, aged 72, but beginning probably, before his seventieth year.

The period during which the greatest number of cases were seen was that between the 30th and 40th years and next, between that of 40 and 50. In cases of bulbar paralysis, the age of 53, seems to be a singularly serious year. I note the occurrence of six cases at about that period of time, amongst 12 in all. There were about five times as many cases among males as among females, but this does not apply to all types of the disease. In the bulbar cases, there were more women, the proportion being 8 to 4.

Amongst the exciting causes, by far the most frequent is that of some form of strenuous occupation. Workmen who had to do very heavy work, athletes or professional contortionists, were among the victims, and in the cases of bulbar paralysis, there was several times a history of the patients being very great and excessive talkers. Lead, as a cause of paralysis, does not seem to me after all, as important a factor as has been supposed. It could be blamed for the disease in not over five cases, and not certainly, in all of these. Several of my patients were very heavy users of tobacco, but I fancy this was only a coincidence. Two patients had in infancy a previous attack of poliomyelitis. In one case there was a very distinct history of an acute infection by dengue; it is not improbable that other cases have followed an acute infection. In one instance, for example, the patient seemed to develop

the trouble after a severe attack of the grip, and it has occurred to me as being not unlikely that the progressive muscular atrophies, which are not due to any specific infection, directly or indirectly, may be due to some degenerative influence of another toxin. In other words, where they may not be parasyphilitic, they are parainfective diseases.

Résumé—All groups of central progressive muscular atrophy not due to tumor, are essentially the same disease, in genesis, course and underlying pathological condition, except that we may separate the progressive occupation atrophies, and certain subacute atrophies. Amyotrophic lateral sclerosis though pathologically and essentially of the character as the other atrophies may receive a separate name on account of its clinical and anatomical peculiarities. The grouping would be then

Progressive Muscular Atrophy (ophthalmoplegic, bulbar, cervical, lumbo-sacral).

Amyotrophic Lateral Sclerosis.

Progressive Occupation Atrophy (not the ordinary limited and arrested type).

Subacute Progressive Atrophy (often recurrent and usually syphilitic).

Progressive Muscular Atrophy is most often excited by prolonged muscular over-exertion. It is in one-quarter, at least, of the cases, parasyphilitic, and in others parainfectious.

APPENDIX.

DUCHENNE-ARAN CASES.

Among 20 cases of the Duchenne-Aran type, there were males 17, females, 3. The ages at onset were:

10-15	3	41-45	1
16-20	1	46-50	0
21-25	2	51-55	4
26-30	1	56-60	1
31-35	2	61-65	0
36-40	4	66-70	1

Total 20

Excessive exertion was noticed in 6.

Excessive use of tobacco in 3.

Syphilis in 8.

In these cases the disease did not reach the bulb while under observation.

Shoulder-Girdle and Hip-Girdle or Root-Type.—This included the cases of so-called chronic occupation atrophies but some were apparently only ordinary progressive muscular atrophy in which the forearms and hands, legs and feet were not involved or but slightly. There were 7 in all, 6 males and ranging in age from 22 to 50, most being between 30 and 50. The distinctly occupation cases ran a much slower course and as stated the trunk muscles became involved. In all but two cases the trouble began in the shoulders and arms. Fibrillary twitchings were present. The knee-jerks were absent where the thighs involved; in other cases exaggerated or normal, but there was no clonus. In none had the bulb become involved. One case was under observation 8, and one 7 years.

SYPHILIS.

The distribution of the syphilitic cases is shown in the table. It is most common in the high palsies, absent in the occupation cases, and in the amyotrophic lateral sclerosis cases. Dr. Collins writes me that he found a syphilitic history in 2 out of 9 cases.

In one case a tabes of specific origin and of long duration ended in bulbar palsy. In one case tabes and bulbar palsy ran together, the bulbar palsy being the dominant thing. In one case tabes and cervico-spinal types ran together.

Syphilis is much the most common in the bulbo-spinal types and all my cases were in men.

HEMIPLEGIC TYPES.

Of the hemiplegic types there were 6 cases. They were 5 males and 1 female. The ages ranged from 26 to 56. There was a history of syphilis in 2 and of lead in 1. The disease began in the leg and then in the arm in 3 cases, in the arm and then in the leg in 2 cases. In 3 cases the trouble was on one side only while under observation. In one case the bulb was finally involved, this being a syphilitic case with absent knee-jerk. In another case not syphilitic the knee-jerks were feeble and the ankle-jerk gone on the affected side. One of these cases presented the history of a subacute or chronic poliomyelitis (see history).

The case had increased knee-jerks but no clonus, and amyotrophic lateral sclerosis was supposed to be developing. Fibrillary tremors and wasting were present in all.

TABULAR SUMMARY.

		Male	Female	With		
				Tabes	Syphilis	Spastic Cases
I	Progressive ophthalmoplegia ending in bulbar spinal atrophy	2	0	0	0	2
	Progressive bulbar palsy, including cases later involving arms and legs ..	10	4	2	4	14
II	Duchenne-Aran cases without later bulbar involvement, ...	27	6		9	5
	With bulbar involvement, ...	11	2		6	2
	Hemiplegic and other types	3	0	0	0	3
III	Typical amyotrophic lateral sclerosis					
IV	Occupation: Dr. Shoul-	6	1		0	0
	derton's type	—	—	—	—	7
	Totals	59	13		19	14
						2